Severe Valvar Pulmonary Stenosis

Successful Surgical Treatment with Hypothermia and Inflow Occlusion In the First Week of Life

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ALTHOUGH ADVANCEMENTS have been made during the past decade in the surgical techniques for the correction of congenital heart lesions, the clinical application of these technical achievements has been largely limited to patients over two years of age, and only meager progress has been made in the surgical management of severe cardiac problems in neonates and small infants.[1] Since approximately 35 per cent of babies born alive with heart anomalies die at less than two months of age and 50 per cent do not survive the first year,[4] it seems obvious that the mortality rate from congenital heart defects will continue to be high until further progress is made in the diagnosis and treatment of such lesions in neonates and small infants.

Approximately 60 per cent of the corrective surgical procedures for neonates and infants require the use of open heart techniques.[1] This does not necessarily mean that cardiac bypass is required, since many of the open heart procedures can be performed satisfactorily by use of hypothermia and/or inflow venous occlusion.[2] In the case here reported a baby with severe valvar pulmonary stenosis had successful correction by open heart operation using inflow occlusion and hypothermia at the age of six days.

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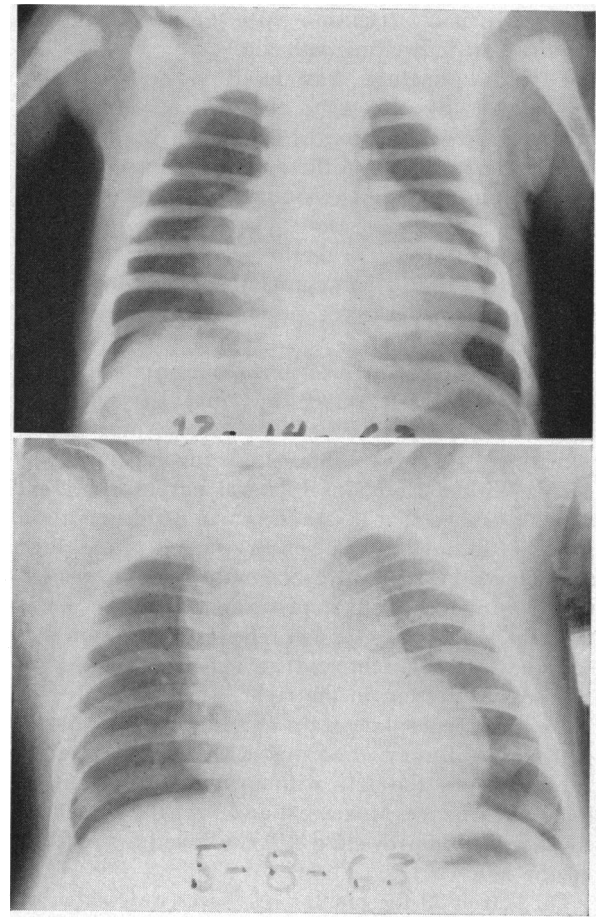

Figure 1.—Preoperative (above) and postoperative (below) roentgenograms of chest (posterior-anterior projection).

Report of a Case

A baby boy two days old was referred to the Sutter Memorial Hospital because of cyanosis, a heart murmur and cardiac enlargement. He was the second child born at term of a 24-year-old mother. The birth weight was 7 pounds 11 ounces. A roentgenogram of the chest taken on the first day of life revealed cardiac enlargement and pronounced pulmonary ischemia. An electrocardiogram had shown right ventricular hypertrophy, probably abnormal for age.

Body temperature was 96° F, the heart rate 120, the respiratory rate 20, blood pressure (flush) 50 mm of mercury in all limbs. There was moderate generalized cyanosis which increased with crying. Slight deformity of the left side of the chest was noted, and there was a thrustful cardiac impulse which was maximal at the xyphoid process. A slight systolic thrill was present at the second left intercostal space along the left sternal border. The first sound was of increased intensity and loudest at the apex. There was fixed splitting of the second sound

and diminished intensity of the second component. A systolic click at the mid left sternal border and a fourth sound at the lower left sternal border were heard. A grade 3 (of six grades) systolic ejection murmur was present at the left base and was well transmitted over the precordium and left posterior chest. There was no diastolic murmur. The liver edge was palpable 4 cm below the right costal margin, but was without pulsation. The peripheral pulses were diminished but were equal in the arms and legs. There was no edema or venous distention.

Hemoglobin was 19.9 gm per 100 ml. Leukocytes numbered 12,600 per cu mm with a normal differential of cells. Results of urinalysis were within normal limits. A roentgenogram of the chest (Figure 1) showed generalized cardiac enlargement and diminished pulmonary vascular markings. An electrocardiogram was interpreted as showing right axis deviation, right atrial hypertrophy and right ventricular hypertrophy, abnormal for age (Figure 2). A phonocardiogram confirmed the auscultatory findings (Figure 3).

It was felt that the most likely diagnosis was severe valvar pulmonary stenosis with an intact ventricular septum and a right-to-left shunt through a patent foramen ovale or a small atrial septal defect. The patient's color did not improve significantly with oxygen therapy. He was digitalized with Lanoxin.® On the fourth hospital day, cardiac catheterization and angiograms were performed. There was a 10 mm (mercury) right atrial "a" wave and a 1 mm mean gradient from the right to the left atrium. The right ventricular pressure was elevated to 140/0/10. The femoral arterial pressure was 90/65. The contour of the right ventricular pressure curve was of pyramid shape (Figure 4). Oxygen saturation of the blood in the femoral artery was 83.1 per cent. The right-to-left at the atrial level was computed to be 35 per cent of the systemic flow. The systemic cardiac index (calculated by the Fick method) was 2.3 liters per minute. Cineangiograms were performed with injections of 75 per cent Hypaque® into the inferior vena cava, the superior vena cava, the left atrial appendage and the right ventricle. A biplane Schonander angiogram was also performed following right ventricular injection of contrast material. The angiographic studies revealed right-to-left shunting of the contrast material across a small atrial defect or patent foramen ovale. The left atrium, left ventricle and aorta appeared normal. There was severe valvar pulmonary stenosis with a central valve opening which was computed to be 2 mm in diameter (approximately 6 per cent of normal for age). The right ventricular outflow tract was widely open during diastole but narrowed decidedly during systole (Figure 5).

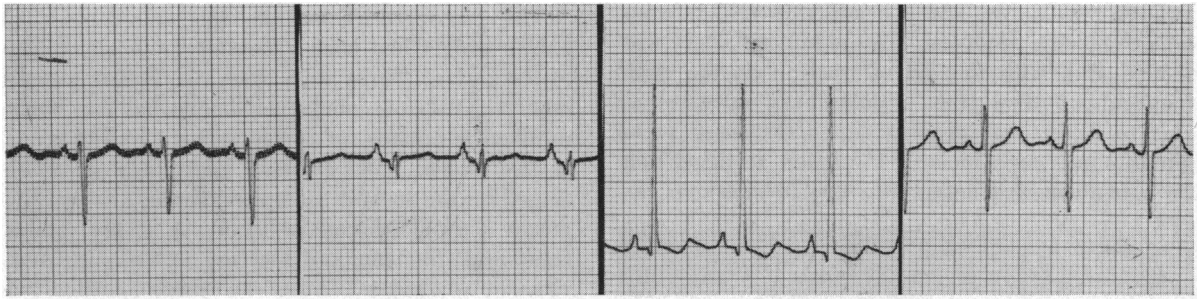

Figure 2.—Preoperative electrocardiograms. From left to right, leads 1, AVF, V_1 and V_6 are shown. Recorded at 25 mm per second.

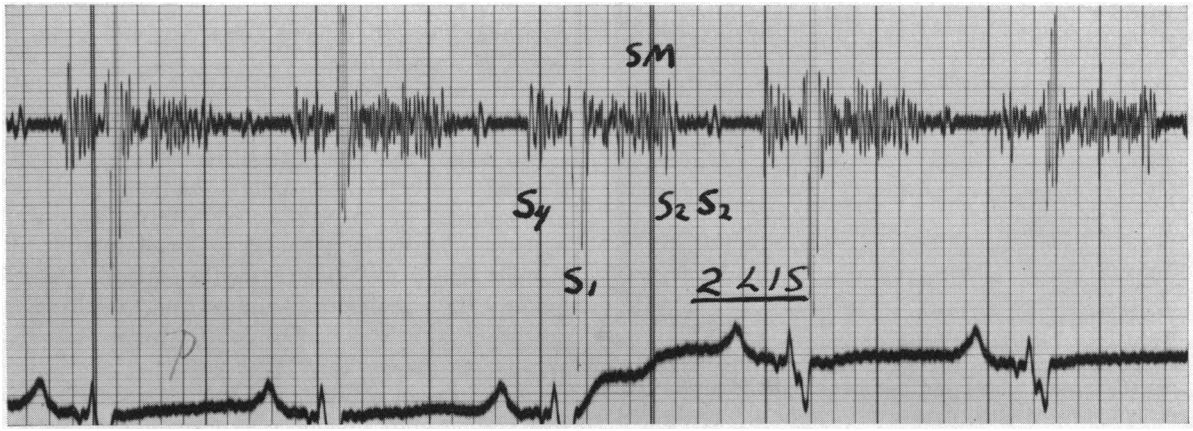

Figure 3.—Preoperative phonocardiographic findings recorded at the left base. Paper speed is 75 mm per second. The electrocardiogram (lead 2) is the reference tracing.

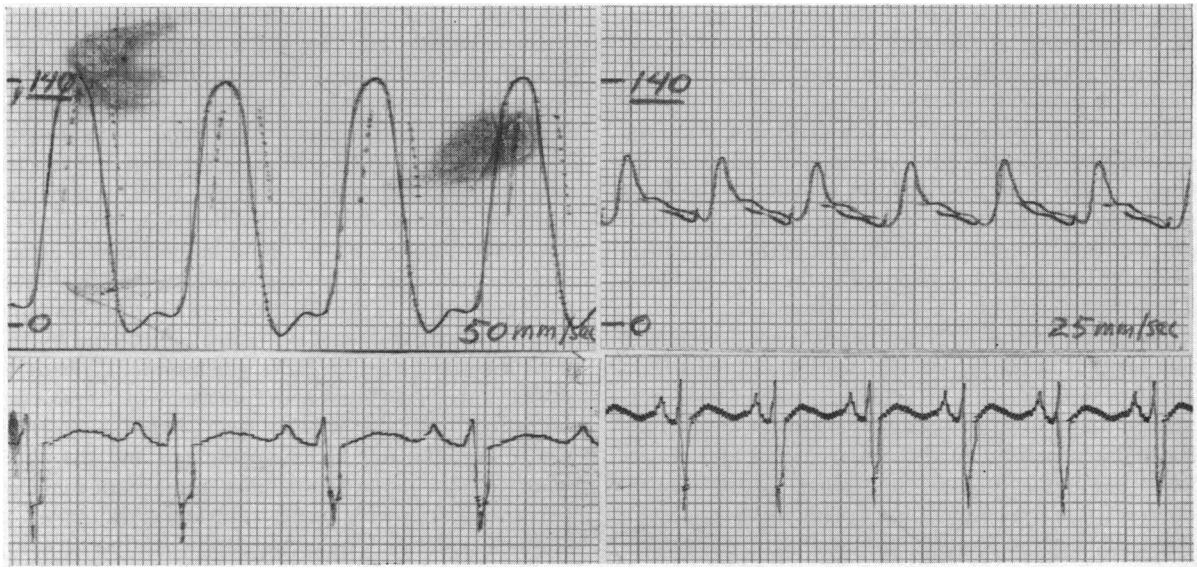

Figure 4.—Right ventricular (above, *left*) and femoral arterial (above, *right*) pressure tracings. The right ventricular systolic pressure of 140 mm of mercury was 50 mm higher than the femoral arterial systolic pressure. The lower frames are corresponding electrocardiograms.

As it appeared the baby would not survive without surgical correction of the right ventricular obstruction, open transarterial pulmonary valvotomy using hypothermia and inflow occlusion was carried out when he was six days old. The body temperature was lowered to 34° C, a longitudinal sternal splitting incision was made, and the superior and inferior vena cava were occluded. The pulmonary artery was opened and, with the aid of traction on the right ventricular outflow tract, a pulmonary valve orifice of 1 to 2 mm was visualized and the fused valve commissures were incised. Palpation revealed that the subvalvar obstruction was due to muscular hypertrophy and not to a fixed infundibular obstruction. The pulmonary artery was closed and the caval ligatures released. Total caval occlusion time was two and a half minutes. Total operation time was 100 minutes. The immediate postoperative right ventricular pressure was 20/0/5.

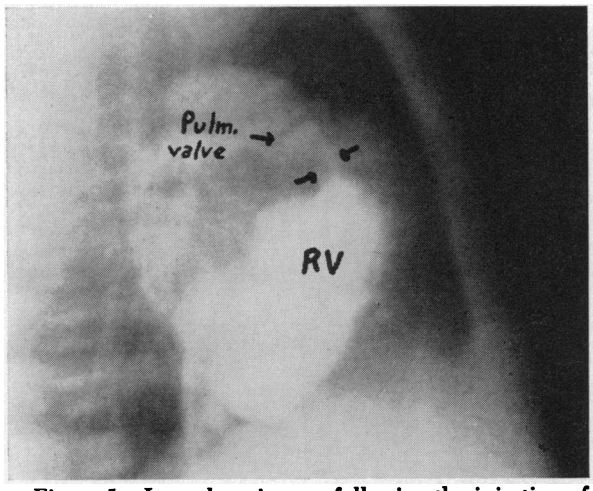

Figure 5.—Lateral angiogram following the injection of contrast material into the right ventricle. The ventricle is in the systole phase and the arrows delineate the pronounced obstruction of the subvalvar outflow tract. During diastole the outflow tract was not obstructed. The dome-shaped, thickened pulmonary valve is also demonstrated.

The postoperative course was uneventful. The baby's weight had been 7 pounds 7 ounces the day of operation. Three days later he began to gain approximately one ounce per day and at four weeks weighed 9 pounds, at seven weeks 12 pounds and at five months 19½ pounds. Digitalis was discontinued on the second postoperative day. Roentgenograms showed progressive decrease in the cardiac transverse diameter and increase in the pulmonary vascular markings (Figure 1). Electrocardiograms continued consistent with right ventricular hypertrophy. At the time of this report the patient was five and a half months old and was asymptomatic. A grade 3 systolic ejection murmur still could be heard at the left base.

Discussion

We believe that open heart correction using hypothermia and inflow occlusion is the best method for management of uncomplicated severe valvar pulmonary stenosis in neonates or small infants. We thought that in the present case the patient's "muscle bound" right ventricular outflow tract[5] might cause persistent right ventricular systolic hypertension of significant degree even after relief of the valvar stenosis.[3] In our experience, older patients who have had angiographic evidence of pronounced obstruction of right ventricular outflow during systole have

all required outflow patching for immediate relief of their right ventricular systolic hypertension. The pressure in the present case dropped to normal immediately following valvotomy. It is possible that the problem of persistent muscular obstruction following valvotomy is related to the duration of right ventricular overwork and consequently does not occur, or is not so frequent, in neonates and young infants.

The case presented also demonstrates the value of cineangiocardiography for the demonstration of stenotic lesions. The pulmonary valve orifice was clearly demonstrated on only two frames of the cineangiogram (60 frames per second). It is, therefore, not unexpected that the valve orifice was not outlined on the Schonander examination when films were taken at a speed of three per second. The size of the pulmonary valve orifice as we calculated it from the angiograms correlated well with the surgical findings. The assessment of the severity of stenotic lesions by the use of positive and negative contrast streams demonstrated cineangiographically will be the subject of a later report.

Summary

Severe valvar pulmonary stenosis in a six-day-old boy was successfully corrected by surgical operation using hypothermia and inflow occlusion. Right ventricular systolic hypertension was immediately relieved in spite of the patient's having a "muscle bound" right ventricular outflow tract.

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Multiple Malignant Lesions With Long-Term Survival

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THE OCCURRENCE of more than one primary malignant growth in one person, while not common, has been reported many times in the medical literature. This is particularly true of primary growths involving multiple sites in a single organ such as the colon. However, the development of three distinct malignant lesions of unrelated cellular type in widely separated areas of the body, as happened in the case here reported, is decidedly unusual. The patient underwent three radical surgical resections of unrelated neoplasms within a period of 19 years. In addition, during this time he also had a severe fulminating episode of pneumococcal meningitis and had six basal cell growths removed from his face.

Report of a Case

Admission 1. A 36-year-old Air Force officer was admitted to an Air Force hospital in Amarillo, Texas, February 3, 1944, with a painful, swollen right testicle. In December, 1943, he had had a contusion of the right groin when he was accidentally kicked while playing volleyball. During the following week the scrotum increased in size on the right and became progressively more tender. The patient was treated as an outpatient for several weeks with cold packs and a scrotal suspensory. When the swelling did not subside and the pain became more intense, he was put in hospital. On admission the

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